BRIEF COMMUNICATIONS

LATE THROMBOSIS OF THE NATIVE AORTIC ROOT AFTER NORWOOD RECONSTRUCTION FOR HYPOPLASTIC LEFT HEART SYNDROME

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Thrombus formation within the native aortic root after Norwood reconstruction for hypoplastic left heart syndrome (HLHS) is theoretically considered to be a lethal condition. This is due to the critical role of the aortic root as a conduit for retrograde blood flow supplying the coronary arterial circulation. We encountered a patient who had undergone a recent extracardiac conduit Fontan operation for completion of staged palliation of HLHS. After a difficult and protracted hospital course, a large, nearly occlusive thrombus within the aortic root was identified by transesophageal echocardiography. Review of her hospital course linked several apparently unrelated clinical events that correlate with thrombus in this location. Surgical thrombectomy was successfully performed and she had an uneventful recovery.

Clinical summary. A 3-year-old, 14-kg girl with HLHS underwent a Norwood operation at 1 week of age involving aortic arch augmentation with a homograft patch. At birth, the ascending aortic diameter was 3 mm, and the aortic valve was severely stenotic with a small amount of antegrade flow. The proximal aortic incision was to the level of the sinotubular junction. She recovered uneventfully and at 7 months of age underwent a bidirectional superior cavopulmonary shunt. She returned at 3 years of age for an extracardiac conduit Fontan operation. Preoperative hemodynamics were excellent by cardiac catheterization, and an 18-mm polytetrafluoroethylene* tube graft was used for Fontan construction. Postrepair transesophageal echocardiography demonstrated excellent ventricular function. Notably, on retrospective review, the aortic root was visualized and no abnormality was present. The aortic root diameter was 10 mm. She had an uneventful recovery and was discharged to her home on a regimen of diuretic and routine warfarin sodium (Coumadin) therapy 2 weeks postoperatively.

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*Gore-Tex graft; registered trade name of W. L. Gore & Associates, Inc, Flagstaff, Ariz.

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She was readmitted 3 weeks later with fatigue and respiratory distress. Large pericardial and pleural effusions were treated with catheter drainage and maximal diuretic therapy. Echocardiography demonstrated decreased ventricular function, which was markedly different from her pre-discharge study. The aortic root was not visualized at this time. By hospital day 12, her ventricular function was good. On hospital day 14 she had transient left hemiparesis while on a sub-therapeutic regimen of warfarin. During this time she had been receiving enoxaparin. Magnetic resonance imaging of the brain showed a small zone of ischemia in the right frontoparietal cortex.

Because of persistent pericardial effusion despite prolonged tube drainage, she underwent creation of a right pleuropericardial window on the 33rd hospital day. The following morning, she had intermittent Wenckebach rhythm and global ST-segment changes, which were transient. Echocardiography showed depressed ventricular function but no other abnormality. The aortic root was not visualized on transthoracic imaging. A large loculated posterior pericardial effusion persisted, and 3 days later she returned to the operating room for a left pleuropericardial window and drainage of the loculated effusion. During this procedure, intraoperative transesophageal echocardiography demonstrated a 7-mm thrombus occupying most of the native aortic root (Fig 1). Given that the age of the thrombus was less than the time period since the Fontan operation, a brief trial of thrombolytic therapy was attempted. After 48 hours of intravenous tissue plasminogen activator failed to reduce the clot size, as determined by transesophageal echocardiography, she was taken to the operating room for surgical thrombectomy. Opening the ascending aorta disclosed a 7-mm soft thrombus occupying the majority of the native aortic root. The thrombus was adherent to the left coronary sinus of Valsalva and propagated into the ostium of the left main coronary artery, which was totally occluded. The clot was extracted with fine forceps and suction, and a left main coronary thrombectomy was performed. The aortic valve was slightly patent and there was no evidence of left ventricular or subaortic thrombus. After weaning from cardiopulmonary bypass, ventricular function was substantially improved on the same amount of dopamine as that before bypass. She was aggressively maintained on anticoagulants postoperatively and had an uncomplicated recovery with resolution of effusions and no further neurologic symptoms or sequelae. Ventricular function was excellent at discharge.

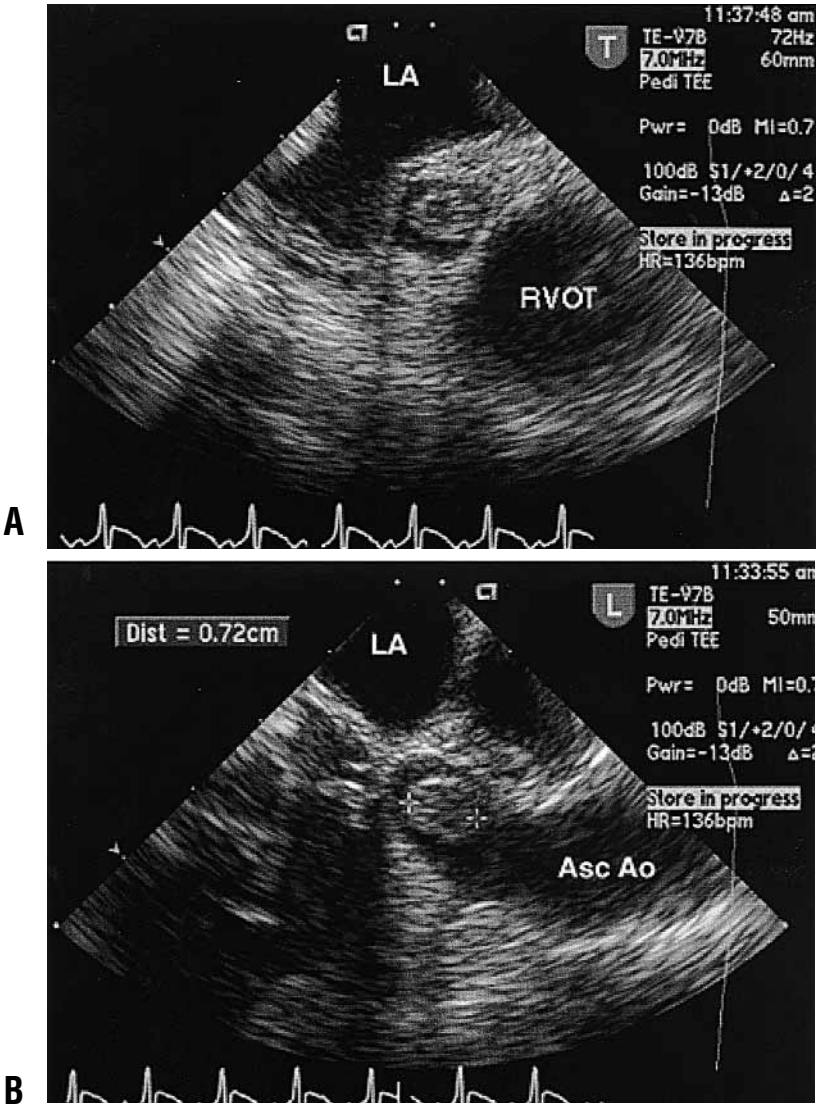

Fig 1. Transesophageal echocardiography images. **A,** Transverse plane at the base of the heart. The native proximal aortic root is seen in cross section immediately anterior to the left atrium. Thrombus occupies most of the lumen. The right ventricular outflow tract is anterior and to the left of the native aorta. **B,** Longitudinal plane. Proximal ascending aorta just above the aortic valve, heading cephalad. On echocardiographic imaging, the thrombus appeared sessile; thrombus movement was most likely restricted by the adjacent aortic walls. *LA,* Left atrium; *RVOT,* right ventricular outflow tract; *Asc Ao,* ascending aorta.

Discussion. Patency of the aortic root after the Norwood operation is critical in that it is the sole conduit for blood flow to the coronary circulation; any compromise can result in hemodynamic instability or death. Because of this concern, it is a surgical priority to minimize the length of the aortic root by constructing the neoaortic arch anastomosis as proximally as possible without compromising the coronary ostia. Suture placement in this region is meticulous to minimize the risk of early or late stenosis of the root orifice. Coronary insufficiency is a major cause of morbidity and mortality after Norwood reconstruction for HLHS. In a postmortem study of 122 patients who had undergone the Norwood procedure, impairment of coronary perfusion was found in 27%, making it the most important cause of death in this series.[1] Of these, 9% died late as a result of chronic myocardial ischemia. The majority had stenosis at the orifice of the aortic root coming off the neoascending aorta. Two patients had thromboembolus in the right coronary artery. Thrombotic occlusion of the left main coronary ostium has been reported in HLHS; however, this was an intraoperative finding during a Norwood operation.[2]

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Survival in a Norwood patient with thrombus in the precoronary aortic root has not been described. In retrospect, this patient's symptoms and protracted clinical course may be explained by thrombus in this location. Her persistent effusions and fatigue may have been due to impaired ventricular function as a direct consequence of subacute coronary insufficiency. The variability in ventricular function, intermittent rhythm change, and electrocardiographic change may also have been related to waxing and waning subacute coronary ischemia or intermittent coronary thromboembolism. The transient ischemic attack is attributable to the presence of thrombus in the ascending aorta; this patient likely had embolic showering from the aortic root into either the systemic or coronary circulation and was at high risk for major thromboembolism. The status of the aortic root in patients late after Norwood construction is not typically sought out during routine evaluation and has not been systematically studied. Our experience with this case argues for careful echocardiographic imaging of the native aortic root in patients with a complicated clinical course after Norwood reconstruction in whom a cause has not been identified.

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